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Case Report

Giant Sinonasal Ameloblastoma

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Abstract

Ameloblastoma is the most common benign odontogenic tumors representing 10% of all tumors that arise in the mandible and maxilla. Ameloblastoma is a slow-growing and locally invasive tumor that presents with painless swelling of maxilla. The diagnosis of ameloblastoma requires CT scan as well as biopsy. The best treatment is surgical. This work reports one giant case of sinonasal ameloblastoma removed by an open surgical procedure.

Keywords: Ameloblastoma; Facial deformity; CT Scan.

Introduction

Ameloblastoma is well recognized as a locally invasive benign neoplasm thought to arise from the cellular components of the enamel organ. Broca described ameloblastoma in 1868 [1]. Ameloblastoma contributes to about 1 % of all head and neck tumors and 13 to 58% of all odontogenic tumors [2]. Maxillary ameloblastoma is rare and account for 15% of all ameloblastomas. Slow-growing, painless swelling of the mandible or maxilla is the most common presentation of ameloblastoma and the diagnosis requires imaging (CT scan). Mutations in genes that belong to the mitogen-activated protien kinase MAPK pathway are found in many ameloblastomas, the most common being the BRAFV600E mutation [3].

Case report

A 68-year-old man consulted in our ENT department with a giant painless mass in the right hemi face since 15 years. This lesion caused major cosmetic and psychological disturbance: isolation and social difficulties. Inspection of face noted major deformity by an ovoid swelling well- limited, non tender (Figure 1) extending to the lower orbital margin and involving the complete right side of the face. The skin was normal without ulceration or inflammation. Intra oral examination was normal. Nasal endoscopy showed a normal mucous and internal displacement of the lateral side of the right nasal cavity. CT scan showed a large heterogenic mass located on the right side of the face, invading the left left maxillary and ethmoidal sinus (Figure 2A,2B). The patient classed stage II (Yang classification). After a discussion with the patient and his family we chose a surgical procedure by external or open approach. A lateral rhinotomy has occurred with a transfixed upper low and right vestibular incision. A large hemi right facial flap was obtained, authorizing a well exposing of the lesion. The mass was removed with a lateral nasal wall The histopathologic study of the surgical specimens confirmed the diagnosis of amelobalstoma without malignancy (Figure 3). Follow- up was full after 8 months, and the patient has recovered social and relational activities (Figure 4).

Discussion

Sinonasal ameloblastomas are rare tumors of the sinonasal tract and show a predilection for the male gender with 59 years mean of age. Maxillary ameloblastoma is more aggressive with a 50% rate of recurrence within 5 years of initial resection [3]. It is generally a painless, slow growing, locally aggressive tumor causing expansion of the cortical bone. The symptoms include deformity (face or palatal deformity), headache, nasal obstruction, epistaxis, intra oral ulceration (palatal ulceration).

The maxillary lesions and extensive lesions require CT and MRI to establish the extent of the lesion. The biopsy confirms the diagnosis and authorize reflection, and adaptable management of the ameloblastoma case with case.

Yang suggested a classification based on diameter of tumor and proposed three stages: stage I, the maximum tumor diameter ≤6 cm; stage II, the maximum diameter of tumor >6 cm or tumor invasion into the maxillary sinus or orbital floor; and stage III, tumor invasion of the skull base or metastasis into regional lymph nodes [4].

According to the new 2022 World Health Organization (WHO) classification of ameloblastomas, they are classified in: unicystic, extraosseous/peripheral, conventional, adenoid and metastasizing ameloblastoma [5].

The histological varieties of ameloblastoma is important to identifying because it was frequently associated with one or multiples recurrences: granular cell ameloblastoma, follicular and plexiform type [6].

The differential diagnosis include inverted papilloma (follicular and acanthomatous ameloblastoma), odontogenic fibromas, non-keratinizing squamous cell carcinoma, adenoid cystic carcinoma, myeloma, sarcoma. The immunohistochemistry may be help full and, all ameloblastoma cell express CK19 (odontogenic epithelium marker) [6].

Recent advances report the detection of mutation in ameloblastomas interesting from newer treatment options. A high incidence of BRAF V600E and SMO L 412F. The oncogenic BRAFV600E mutation leads to the activation of mitogen-activated protein kinase (MAPK) pathway, which has resulted in successful treatment with BRAF inhibitor [7].

The challenge in managing ameloblastoma is in achieving complete excision and reconstruction of the defect when the tumor is large (Table 1).

Surgical resection is treatment of choice. Radical resection with a margin of a least 1 to 2 cm is ideal to obtain save results. Furthermore the radical treatment strategy is associated with a higher risk of post operative complications and required numerous surgical operation (recurrence) and prosthetic procedures. The quality of life of patients is significantly altered with pain and local deformity. A conservative treatment, curettage, has a recurrence rate of 60 to 80% [3,5].

Endoscopic sinus surgery can to be used in some selected cases (karp). In 2021 Karp report only 2 cases in the literature of endoscopic resection of ameloblastoma with respectively 4-year and 11 months follow-up [8,9].

Non-surgical treatment in ameloblastoma comported systemic chemotherapy (metastatic ameloblastoma) especially platinum-based anticancer molecular. Recently molecular targeted therapy was cited in many works: vemurafenib, dabrafenib and trametinib showed a notable reduction in tumor volume [10].

Radiotherapy is utilized in select cases like residual disease after surgery, multiple recurrences, impossibility of surgery or unresectable lesions (66 to 70 Grays) [11].

The prognosis for ameloblastoma depends on the age of the patient, location and size of the tumor, histological type, extent, and stage of disease. The recurrence rate of 9,8% to 19,3% after treatment, and more than 50% of recurrences occur within five years of the primary surgical intervention.

Table 1: Ameloblastoma protocol of management.

Section	Modalities
Diagnosis	Imaging: CT Scan. Biopsy (accessible lesion).
Evaluation and Staging	CT scan MRI Staging (Yang classification)
Therapeutic Protocol	Surgical option Radical surgical resection with margin (1.5-2 cm). Reconstruction: Flaps or Prosthetic reparation
Post Operative Evaluation and Staging	Histopathologic study -Histological type -Quality of resection: margin Identification of prognosis factory -Age -Histological variety -Extension: base of skull, orbit, cerebral -Recurrence -Maxillary or soft tissues extension
Follow-Up	CT Scan Every 6 months during 2 years.
Ameloblastoma with Poor Prognosis	Research of Mutation Braf V600E
Recurrence 1 OR 2	Gold Option Surgical Protocol (Radical Resection).
Multiples Recurrences	Gold option: surgical protocol Option 2: radiotherapy Option 3: targeted therapy
Metastatic Ameloblastoma	Evaluation: TEP Chemotherapy Targeted therapy

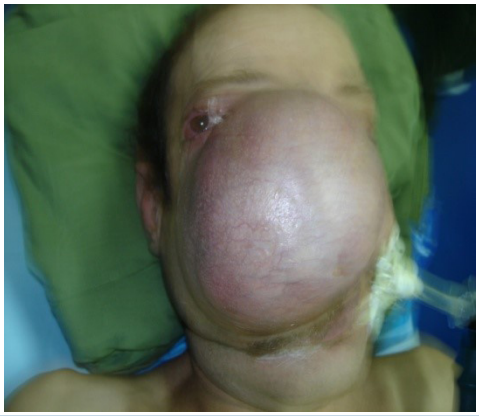

Figure 1: Clinical presentation- giant facial deformity mass on the right side.

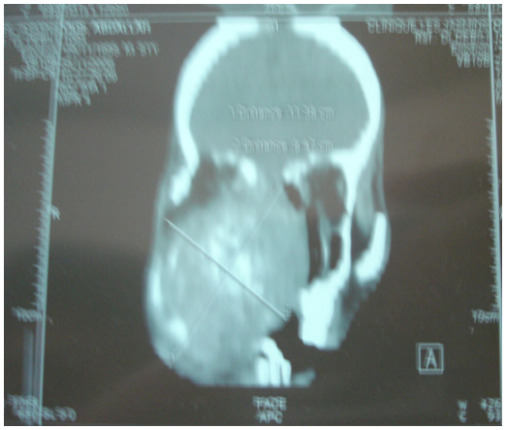

Figure 2a: CT scan coronal view- voluminous heterogeneous mass on the right side of face involving maxillary sinus.

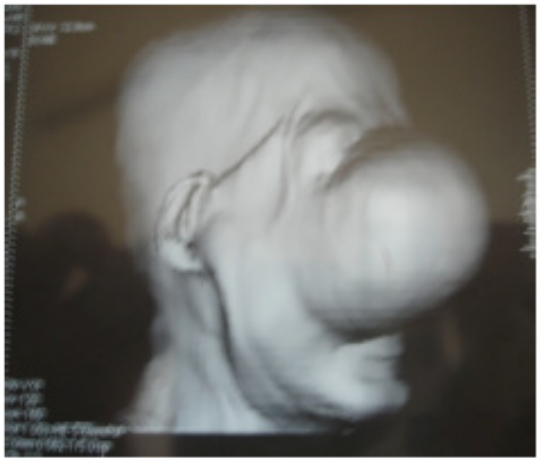

Figure 2b: 3D-CT scan voluminous heterogeneous mass on the right side of face.

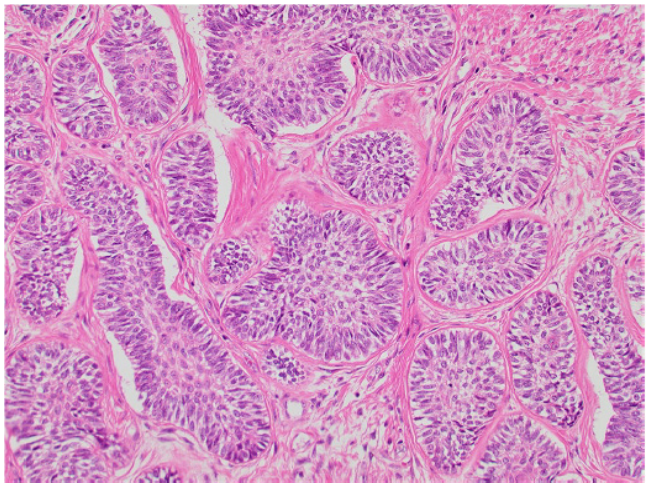

Figure 3: Histopathology - Follicular patern.

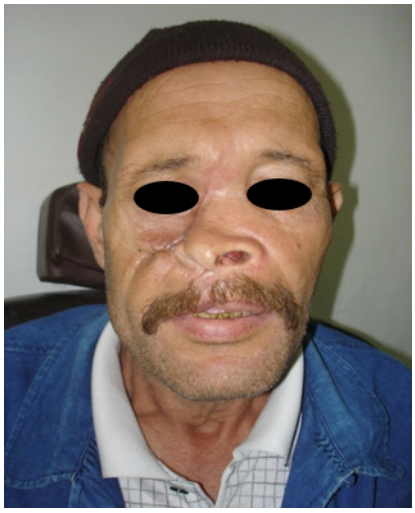

Figure 4: Post-operative results after removing the tumor.

Conclusion

Ameloblastoma, an odontogenic tumor variety is rare with a locally invasive potential, slow-growing with painless swelling and deformity. CT scan shows the lesion, location and extension. The best treatment of ameloblastoma is surgical especially in bloc resection (radical option). The future may be based on molecular developments, with the possibility of targeted therapy.

References

1. Martin Y, Sathyakumar M, Premkumar J, Magesh KT. Granular cell ameloblastoma. J Oral Maxillofac Pathol. 2017; 21(1): 183. doi: 10.4103/jomfp.JOMFP_45_15.
2. Fregnani ER, da Cruz Perez DE, de Almeida OP, Kowalski LP, Soares FA, de Abreu Alves F. Clinicopathological study and treatment outcomes of 121 cases of ameloblastomas. Int J Oral Maxillofac Surg [Internet]. 2010; 39(2): 145-9.
3. Sweeney RT, Mc Clary AC, Myers BR, Biscocho J, NeahringL, Kwei KA. Identification of recurrent SMO and BRAF mutations in ameloblastomas. Nat Genet. 46: 722-725. DOI 10.1038/hg.2986.
4. Yang R, Liu Z, Gokavarapu S, Peng C, Ji T, Cao W. Recurrence and cancerization of ameloblastoma: multivariate analysis of 87 recurrent craniofacial ameloblastoma to assess risk factors associated with early recurrence and secondary ameloblastic carcinoma. Chin J Cancer Res. 2017: 189-95. 10.21147/j.issn.1000-9604.2017.03.04.
5. Soluk-Tekkesin M, John M, Wright JM. The World Health Organization Classification of Odontogenic Lesions: A Summary of the Changes of the 2022 (5th) Edition. Turk Patoloji Derg. 2022; 38(2): 168-184. doi: 10.5146/tjpath.2022.01573.
6. Ghai S. Ameloblastoma: An updated narrative review of an enigmatic tumor. Cureus. 2022; 14(8): 27734. doi: 10.7759/cureus.27734.eCollection2022 Aug.
7. Kurppa KJ, Caton J, Morgan PR, et al. High frequency of BRAF V600E mutations in ameloblastoma J Pathol 2014; 232: 492 -8.
8. Karp J, Xiong W, Derikvand S, Javer A. Maxillary Sinus Ameloblastoma: Transnasal Endoscopic Management. Ear Nose Throat J. 2021; 100(10): 908S-912S. doi: 10.1177/0145561320930555.
9. Lee J, Ahmad ZA, Kim D, et al. Comparison between endoscopic prelacrimal medial mxillectomy and caldwell-Luc approach for benign maxillary sinus tumors. Clin Exp Otorhinolaryngol 2019; 12(3): 287-293.
10. Kaye FJ, Ivey AM, Drane WE, Mendenhall WM, Allan RW. Clinical and radiographic response with combined BRAF-targeted therapy in stage 4 ameloblastoma. J Natl Cancer Inst. 2015; 107: 378. 10.1093/jnci/dju378.
11. Koukourakis GV, Miliadou A, Sotiropoulou-Lontou A. Ameloblastoma, a rare benign odontogenic tumour: an interesting tumour review targeting the role of radiation therapy. Clin Transl Oncol. 2011; 13: 793-7. 10.1007/s12094-011-0735-5.